Page 5, replace the sixth paragraph starting on line 29 with:

G4

Cosmids were from a cosmid library constructed by subcloning YAC 774G4 (Richard et al., 1995) and are presented as lines. Dots on lines indicate positive STSs (indicated in boxed rectangles). A minimum of three cosmids cover the entire gene.

Page 6, replace the first paragraph starting on line 1 with:

G5

Figures 2A-2C: Sequence of the human nCL1 cDNA (B), and the flanking 5' (A) and 3' (C) genomic regions.

A) (SEQ ID NO:68) and C) (SEQ ID NO:69) The polyadenylation signal and putative CAAT, TATAA sites are boxed. Putative Sp1 (position -477 to -472), MEF2 binding sites (-364 to -343) and CArG box (-685 to -672) are in bold. The Alu sequence present in the 5' region is underlined.

B) The corresponding amino acids are shown below the sequence. The coding sequence between the ATG initiation codon and the TGA stop codon is 2466 bp (SEQ ID NO:70), encoding for an 821 amino acid protein (SEQ ID NO:6). The adenine in the first methionine codon has been assigned position 1. Locations of introns within the nCL1 gene are indicated by arrowheads. Nucleotides which differ from the previously published ones are indicated by asterisks.

Page 6, replace the third paragraph starting on line 24 with:

G6

Figures 4A-4B: Distribution of the mutations along nCL1 protein structure.

Page 7, replace the third paragraph starting on line 9 with:

G7

Figures 7A-7D: Homozygous mutations in the nCL1 gene

Page 7, replace the fourth paragraph starting on line 15 with:

G8

Figures 8A-8D: Structure of the nCL1 gene

Page 11, replace the second paragraph starting on line 3 with:

Table 2: Sequences at the intron-exon junctions (SEQ ID NO:71-SEQ ID NO:116). A score expressing adherence to the consensus was calculated for each site according to Shapiro and Senapathy (1987). Sequences of exons and introns are in upper and lower cases, respectively. Size of exons are given in parentheses.

G9

Page 16, replace the sixth paragraph starting on line 30 with:

As expected, due to multiple consanguineous links, the examined LGMD2A Northern Indiana Amish patients were homozygous for the haplotype on the chromosome bearing the mutant allele (Allamand et al., 1995). A (G->A) missense mutation was identified at nucleotide 2306 within exon 22 (Fig. 7). The resulting codon change is CGG to GAG, transforming Arg^{769} to glutamine. This residue, which is conserved throughout all members of the calpain family in all species, is located in domain IV of the protein within the 3rd EF-hand at the helix-loop junction. This mutation was encountered in a homozygous state in all patients from 12 chromosome 15-linked Amish families, in agreement with the haplotype analysis. We also screened six Southern Indiana Amish LGMD families, for which the chromosome 15 locus was excluded by linkage analyses (Allamand ESHG, submitted, ASHG 94). As expected, this nucleotide change was not present in any of the patients from these families, thus confirming the genetic heterogeneity of this disease in this genetically related isolate.

G10

Page 24, before the first paragraph starting on line 2, insert the new paragraph:

Allamand, V., Broux, O., Richard, I., Fougerousse, F., Chiannilkuchai, Bourg, N., Brenguier, L., Devaud, C., Pasturaud, P., Pereira de Souza, A., Roudaut, C., Tiscfield, J. A., Connealy, P. M., Fardeau, M., Cohen, D., Jackson, C. E. and Beckmann, J. S. (1995). Preferential localization of the limb girdle muscular distrophy type 2A gene in the proximal part of a 1-cM 15q15.1-q15.3 interval. Am. J. Hum. Genet. *56*,1417-1430.

G11

Page 28, after the first paragraph starting on line 3, insert the new paragraph: